CHRONIC WASTING DISEASE FACT SHEET

May 31, 2013

What is Chronic Wasting Disease?

Chronic wasting disease (CWD) is a fatal, transmissible spongiform encephalopathy (TSE) of cervids. TSEs are caused by abnormally folded forms of a normal prion protein that occurs naturally in the brain of many classes of animals including deer. By unknown mechanisms, the abnormal prion can enter the brain and induce normal proteins to develop the abnormal conformation. These abnormal proteins continue to grow in quantity until they eventually cause lesions in the brain. Similar diseases occur in sheep and goats (scrapie), cattle (bovine spongiform encephalopathy also known as "Mad Cow Disease"), and humans (Creutzfeldt-Jakob disease). Although unconfirmed, CWD may have originated from scrapie in domestic sheep (Williams 2005), and it is the only TSE known to occur in free-ranging wildlife.

What Species are Susceptible to CWD?

Natural CWD infections have been diagnosed in white-tailed deer (*Odocoileus virginianus*), mule deer (*Odocoileus hemionus*), elk (*Cervus canadensis*), red deer (*Cervus elaphus*), Sika deer (*Cervus nippon*), moose (*Alces alces*), and associated subspecies and hybrids. Experimental oral transmission resulted in disease in reindeer (*Rangifer tarandus*) (Mitchell at al. 2012). These susceptible species represent both major cervid subfamilies (Capreolinae and Cervinae). Other members of the cervid family may be vulnerable (Saunders et al. 2012); however, experimental studies are largely lacking. Further, different strains of CWD have been identified, and variation among cervid species in incubation time and/or susceptibility to various strains is unknown. So far, experimental studies have shown that certain genetic varieties (genotypes) of some cervid species, such as elk, mule deer, and white-tailed deer, differ in the time it takes from experimental inoculation to the development of clinical disease, but all genotypes in these species have been susceptible; no true resistance has been observed (reviewed by Robinson et al. 2012).

Interspecies transmission of CWD to non-cervids has not been documented under natural conditions. With the emergence of new CWD strains, however, the potential for infection of humans and other mammals remains (Saunders et al. 2012). Host range and pathogenicity differ among prion strains, and therefore the potential for transmission to other species may vary. Further, transmission of CWD to other mammals (including some primates) has been achieved through experimental intracerebral (into the brain) inoculation (Saunders et al. 2012). Nevertheless, based on epidemiologic studies, the risk for human transmission is believed to be low and such transmission has not been documented (Belay et al. 2004, Wilson et al. 2012).

How is CWD Transmitted?

The exact mechanism of CWD transmission is unknown; however, CWD has been shown to be transmitted directly through animal-to-animal contact and indirectly through contaminated environment (Williams 2005). The environment can become contaminated by decomposing carcasses and through an infected cervid shedding the CWD agent in urine, feces, saliva, blood and antler velvet (Angers et al. 2009, Gough and Maddison 2010). Prions are extremely resistant to degradation and can remain infectious in soil for years. Since cervids normally ingest a considerable amount of soil while foraging, soil has been suggested to play a key role in CWD transmission (Schramm et al. 2006, Smith et al. 2011). Aerosol transmission of CWD in white-tailed deer has also been demonstrated experimentally using an inoculum less than 1/20th of the dose used in oral inoculation trials (Denkers et al. 2012).

Clinical Signs and Pathology

CWD is invariably fatal once the deer is infected. The incubation period (time between infection and onset of clinical signs) varies with host species, host genetics (and other factors), and prion strain but is usually at least 16 months and averages 2 to 4 yrs (Williams 2005). Typically, CWD is characterized by weight loss and behavioral changes occurring over weeks to months. Additionally, neurologic signs such as ataxia (incoordination), head tremors, and wide-based stance are commonly seen (Williams 2005). Death usually occurs within 4 months of the onset of clinical signs, although some animals may survive for up to a year (Williams 2005). Clinical signs are often more subtle and incubation periods longer in elk versus deer (Williams and Miller 2002).

How is CWD Diagnosed?

Early after infection, and long before the onset of clinical signs, the CWD prion can be detected in some lymph nodes and brainstem. These lymph nodes and the brainstem can only be collected postmortem and are tested using a method called immunohistochemistry (IHC). IHC is the only USDA-APHIS-approved testing method for the CWD Herd Certification Program (for captive cervids) (USDA 2012). Enzyme-linked immunosorbent assay (ELISA) testing of these tissues also is an approved method for surveillance in free-ranging cervids.

Currently, there is no USDA-APHIS-approved live-animal test for CWD. Biopsy of the tonsils and rectal mucosa are two promising methods that have been used in live animals. In deer, tonsil biopsy has been used successfully to detect CWD (Wild et al. 2002), however, in elk it may fail to detect early infections (Monello et al. 2013). The sensitivity of rectal biopsies varies with the age and genetics of the animal and, importantly, can miss animals, especially in the early stages of infection (Thomsen et al. 2012, Monello et al. 2013).

Tests for CWD currently are unable to detect the disease in the earliest stages of infection; therefore, no test can guarantee that an animal was uninfected at the time of sampling.

CWD Distribution and Prevalence

CWD was first recognized as a syndrome in captive mule deer in Colorado in 1967 (Williams and Young 1980) but may have been present in free-ranging mule deer in Wyoming and Colorado for more than 50 years (Miller et al. 2000). The disease spread naturally among wild cervids within this area but is believed to have spread to other states primarily via movement of infected captive cervids. In 1996, CWD was first reported outside the "endemic zone" of Wyoming and Colorado in a captive elk in Canada. CWD was found in wild deer in Wisconsin in 2002, the first time east of the Mississippi – prompting an intensive surveillance effort throughout the US and Canada. Since then, additional CWD-positive areas have been identified and new outbreaks have occurred. As of May 2013, CWD has been identified in free-ranging cervids in 18 states and 2 Canadian provinces and in approximately 100 captive herds in 13 states and 2 provinces (Saunders et al. 2012, CWD Alliance website).

In almost all areas where CWD has been diagnosed, the incidence and distribution of the disease appears to be increasing. There are likely multiple causes for the spread of CWD. Locally, and even regionally, the disease appears to spread naturally through the movement and dispersal of infected deer, and possibly through scavengers feeding on infected carcasses. The transport of infected hunter-killed carcasses may also be a mechanism for the spread of CWD. Movement of infected captive cervids, however, is believed to be the most important cause for the spread of CWD in North America. Miller and Williams (2004) stated "[t]he geographic extent of endemic CWD in free-ranging wildlife was initially thought to be quite limited and its natural rate of expansion slow; however, recent investigations have revealed that CWD has been inadvertently spread much more widely via market-driven movements of infected, farmed elk and deer."

The risk of CWD is greater in captive herds because of the high deer density (resulting in both increased contact and higher concentration of prion in soil) and frequent exchange of animals among facilities. Captive cervid escapes or releases from captive facilities in Florida and the rest of the US are not uncommon, and visitation and nose-to-nose contact between captive and wild deer occur regularly – posing a risk for CWD (and other disease) transmission. Even in the absence of direct contact (e.g. double-fence), transmission possibly may occur via aerosolized dust particles and movement of scavengers feeding on infected captive cervid carcasses. With the latter, the CWD prion is able to pass intact through the digestive tract of crows (VerCauteren et al. 2012), and presumably other scavengers, and therefore can potentially be translocated by the scavenger outside of a captive facility.

Despite monitoring and management programs, CWD continues to spread among captive facilities. In the last 10 years, CWD has been diagnosed in captive herds in New York, Michigan, Missouri, Pennsylvania, and Iowa. Many of these herds had been monitored for CWD more than 5 years.

Impact to Free-ranging Populations

The long term impact of CWD on free-ranging populations is unknown. The disease, however, does not appear to stabilize or diminish over time as prevalence and distribution continue to increase. Most early scientific models developed to better understand transmission and population impacts examined direct (deer-to-deer) transmission; however, models including indirect transmission and long-term prion persistence in the environment may be more accurate. These models suggest severe impacts to free-ranging populations (Almberg et al. 2011). Indeed, CWD has been associated with large declines in the mule deer population in areas of Colorado (Saunders et al. 2012, Miller et al. 2008). In Florida, a substantial decline in deer numbers due to the disease and/or management efforts would be a significant

impact to hunting and wildlife viewing opportunities and could affect prey availability for Florida panthers.

Impact to Captive Populations

In captive herds, the process of transmission may be rapidly accelerated, and in the absence of culling, prevalences can approach 100% and lead to complete herd destruction (Williams 2005, Saunders et al. 2012). The initial response to CWD detection in a captive cervid herd is quarantine, followed by extensive investigations of herds that shipped to or received animals from the affected herd. These herds also may be quarantined with no animal movement allowed in or out of the herd. Depopulation of all animals in an affected herd is the preferred response to control the spread of CWD in captive deer and elk. However, federal funds for indemnification are no longer available and herd owners bear the expense if this method is chosen.

Management of CWD

Management strategies to eradicate or even control CWD in wild deer in endemic areas have focused on reducing prevalence and controlling spread by population reduction, culling of suspect or exposed animals, and prohibition of practices, such as baiting and feeding, that artificially concentrate wild cervids. These efforts have heretofore been unsuccessful. Outside of the endemic area, management has focused on early detection followed by deer population reduction. These too, for the most part, have been unsuccessful. Intensive culling of wild populations has failed to halt the spread or decrease the prevalence of CWD in Wisconsin, Illinois, and West Virginia. Only New York, which detected 2 infected wild and 2 captive deer in 2005, has had no recurrence of the disease. Depopulation is likely only effective very early following the introduction of CWD to free-ranging deer, and it often is not feasible. Culling as a management tool becomes even less effective, however, as CWD spreads, prevalence rates increase, and the environmental build-up of infectious prion worsens over time (Almberg et al. 2011).

For captive deer and elk, management recommendations are complete depopulation, extensive follow-up and testing of any cervids that have come in contact (through intra-or interstate movement) with the infected herd, rigorous prevention of any contact with free-ranging deer, and disinfection of all exposed areas (Saunders et al. 2012). However, even with the most stringent decontamination measures, risk of disease recurrence remains (Saunders et al. 2012) at the premises and repopulation with cervids is prohibited or discouraged, as is allowing wild cervids to become exposed to the site.

Prevention of CWD

There is no vaccine or treatment, and eradication or even management of the disease, has largely been ineffective. Therefore, prevention of CWD introduction is the most important method of control. In Florida, prevention measures have included public education and regulations. To address the risk posed by infected hunter-killed carcasses from infected areas, a rule banning the importation of hunter-killed carcasses (except boned-out meat, clean skull cap and antlers, and tanned hides) from CWD-positive states was implemented in 2005. More importantly, following detection of CWD in wild deer in Wisconsin in 2002, an emergency rule restricting the importation of cervids into Florida was implemented by both the Florida Fish and Wildlife Conservation Commission and the Florida Department of Agriculture and Consumer Services (FDACS). This was later replaced by a FDACS rule banning cervid importation except from captive herds that were certified as having no CWD positive animals for ≥5 yrs. This temporarily eliminated legal importation into Florida for several years until herds began to attain the certified status. However, given the continued detection of CWD in captive cervids, the effectiveness of

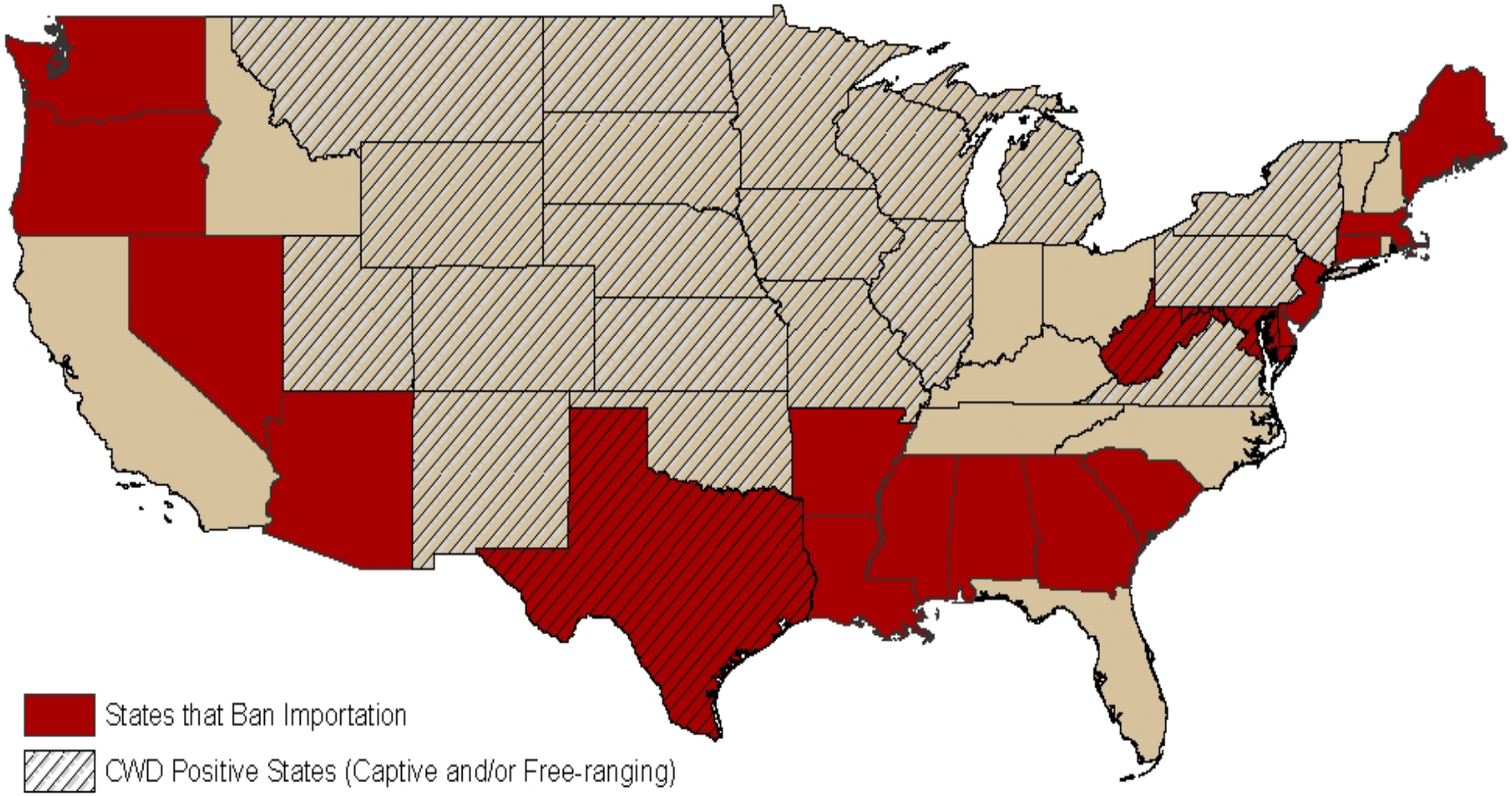
CWD monitoring to establish captive herds as “low risk” for CWD is questionable, and certified status does not guarantee a herd is free from CWD.

In recognition of the risks that captive herds pose for the introduction of CWD and other infectious diseases to wild deer, 16 states have banned the importation of some or all species of cervids into their state, and several states ban captive cervid farms altogether (Figure 1).

Why do we need to do more?

Currently, the closest CWD-positive state to Florida is Virginia. This “buffer zone” of CWD-free states near Florida ensures that any natural movement of a CWD-positive cervid into Florida is highly unlikely. Furthermore, all of our neighboring states ban importation of cervids and therefore create an additional barrier to CWD introduction to Florida. All of this geographic protection, however, is futile as long as we continue to allow cervid importation into Florida. History has shown that movement of captive cervids is likely the greatest risk factor for introduction of CWD into new areas. It is also known that once CWD is present, it is nearly impossible to eradicate, and although the long-term effects of the disease are not known, once established, substantial negative impact to the population is inevitable. Prohibiting the importation of live cervids is the best way to reduce the chances of this disease entering our state and thereby to protect our wild deer population from CWD, help ensure our hunting heritage and protect Florida’s captive deer industry.

Figure 1. Chronic wasting disease positive states and states that ban importation of cervids in the lower 48 states.



* Texas: Ban on all cervid importation except elk
* West Virginia: Department Natural Resources bans all importation, Department of Agriculture prohibits importation of cervids from CWD positive counties
* Nevada and Washington: No captive cervid facilities allowed

Source: CWD Alliance